

B6.129S-Idua ^{tm1.1Kmke} /J

Stock No: **017681** | Idua-W392X

 Congenic, Targeted Mutation

Live mice available in varying quantities. Ask Customer Service for details.

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commonly found in patients with Hurler syndrome and are valuable for evaluation of therapeutic treatments mucopolysaccharidosis type I-Hurler (MPS IH).

Donating Investigator

Kim M. Keeling, University of Alabama at Birmingham

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GENETIC OVERVIEW

Genetic Background

Generation

[N8pN1+N2F3](#)
(2021-04-06 00:00:00)

Idua^{tm1.1Kmke}

Alele Type

Gene Symbol

Gene Name

Targeted

Idua

iduronidase, alpha-L

VIEW GENETICS

RESEARCH APPLICATIONS

Developmental Biology Research
Internal/Organ Research

VIEW ALL RESEARCH APPLICATIONS

BASE PRICE

Starting at:

\$236.78 Domestic price for female

473.55 Domestic price for breeder pair

V I E W P R I C E L I S T

Details

Detailed Description

These mice carry a nonsense mutation at *Idua* codon W392, (*Idua*-W392X), which corresponds to the human IDUA-W402X mutation that prematurely terminates protein synthesis and is commonly found in patients with mucopolysaccharidosis type I-Hurler (MPS IH) syndrome. Deficiency of the alpha-L-iduronidase enzyme causes this lysosomal storage disease. No alpha-L-iduronidase activity is detected in brain and liver tissues from homozygous mice aged 5, 10 and 30 weeks. Although mice that are homozygous for this mutation are viable and fertile, they have a median lifespan of 69 weeks. Homozygotes exhibit a progressive increase in urinary excretion of glycosaminoglycans (GAGs), and progressive accumulation of GAGs in tissues. The level of steady state *Idua* mRNA is reduced 30-50%. Histological analysis reveals progressive accumulation of lysosomal storage inclusions in the cytoplasm of Purkinje cells, medullar neurons, as well as infiltration of foamy macrophages that increases with age. Thickening of the zygomatic arch and femur is detected at 15 weeks of age by radiography, and increases at 35 weeks of age. At 35 weeks of age femur bone mineral density is increased and percent body fat decreased.

Development

Control Suggestions

Selected References

Genetics

Idua^{tm1.1Kmke}

Disease/Phenotype

[+ Disease Terms](#)

[+ Research Areas By Phenotype](#)

[+ Mammalian Phenotype Terms by Genotype](#)

[+ References](#)

[- Technical Support](#)

C O N T A C T T E C H N I C A L S U P P O R T

Genotyping Protocols

Standard PCR:[Iduaalternate1](#)

Probe:[Idua Probe](#)

[Genotyping resources and troubleshooting](#)

Dietary Information

LabDiet® 5K52 formulation (6% fat)

Breeding Considerations

When maintaining a live colony, these mice can be bred as homozygotes; however, homozygotes have a median lifespan of 69 weeks.

[Additional Breeding and Husbandry Support](#)

Mating System

Homozygote x Heterozygote

Heterozygote x Homozygote

Citation

When using the Idua-W392X mouse strain in a publication, please [cite the originating article\(s\)](#) and include JAX stock #017681 in your Materials and Methods section.

Animal Health Reports

[Facility Barrier Level Descriptions](#)

 [FGB29 \(Standard\)](#)

[- Pricing & Availability](#)



Available

Live mice available in varying quantities. Ask Customer Service for details.

Domestic International

Pricing effective for USA, Canada and Mexico shipping destinations

LIVE MOUSE			
AGE	SEX	GENOTYPE	PRICE
Approx 4-8 weeks	Female	Heterozygous for <i>Idua</i> ^{tm1.1Knrke}	\$236.78
	Male	Heterozygous for <i>Idua</i> ^{tm1.1Knrke}	\$236.78
Approx 4-8 weeks	Female	Homozygous for <i>Idua</i> ^{tm1.1Knrke}	\$236.78
	Male	Homozygous for <i>Idua</i> ^{tm1.1Knrke}	\$236.78

BREEDER PAIR			PRICE
SEX	GENOTYPE		
Female	Heterozygous for <i>Idua</i> ^{tm1.1Knrke}		\$473.55
Male	Homozygous for <i>Idua</i> ^{tm1.1Knrke}		
Female	Homozygous for <i>Idua</i> ^{tm1.1Knrke}		\$473.55
Male	Heterozygous for <i>Idua</i> ^{tm1.1Knrke}		

RELATED PRODUCTS AND SERVICES		
Frozen Mouse Embryo	B6.129S- <i>Idua</i> < <i>tm1.1Knrke</i> >/J	\$2595.00

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Terms are granted by individual review and stated on the customer invoice(s) and account statement. These transactions are payable in U.S. currency within the granted terms. Payment for services, products, shipping containers, and shipping costs that are rendered are expected within the payment terms indicated on the invoice or stated by contract. Invoices and account balances in arrears of stated terms may result in The Jackson Laboratory pursuing collection activities including but not limited to outside agencies and court filings.

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The Jackson Laboratory has rigorous genetic quality control and mutant gene genotyping programs to ensure the genetic background of JAX® Mice strains as well as the genotypes of strains with identified molecular mutations. JAX® Mice strains are only made available to researchers after meeting our standards. However, the phenotype of each strain may not be fully characterized and/or captured in the strain data sheets. **Therefore, we cannot guarantee a strain's phenotype will meet all expectations.** To ensure that JAX® Mice will meet the needs of individual research projects or when requesting a strain that is new to your research, we suggest ordering and performing tests on a small number of mice to determine suitability for your particular project. We do not guarantee [breeding performance](#) and therefore suggest that investigators order more than one breeding pair to avoid delays in their research.

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Q U E S T I O N S A B O U T T E R M S O F U S E

ADDITIONAL USE RESTRICTIONS APPLY

Use of MICE by companies or for-profit entities requires a license prior to shipping.

LICENSING INFORMATION

Phone: 207-288-6470

Email: TechTran@jax.org

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By Gene

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